Systemic Lupus Erythematosus: Challenges Tracking a Complex, Multi-Systemic Chronic Disease

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Outline

I. Introduction
II. Purpose
III. Estimating SLE Prevalence in Boston
IV. Linkage Demonstration Project
V. Linkage Results
VI. Summary of EPHT Experience

II. Purpose

- Estimate prevalence & annual incidence rates of SLE in Boston.
- To link SLE cases to environmental data.
- Discuss the limitations & lessons learned.
- Identify future surveillance methods to consider.

Systemic Lupus Erythematosus (SLE)

- Chronic inflammatory autoimmune disorder
- Multisystemic with variety of manifestations:
    - skin
    - joints
    - blood cells
    - heart
    - lungs
    - kidneys
    - nervous system
- Three patterns:
    - Remitting relapsing
    - Chronic active
    - Long quiescence
- Prognosis ranges from mild to fatal.
    - 5 year Mortality 5-10 %

SLE: Gender, Race and Age

- Gender:
  - 5 to 14 times higher incidence in females than males
- Race:
  - 3-6 times higher incidence in Afr-Am or Afr-Carib than Caucasian
  - Other races seem higher than Caucasian but less studied
  - Afr-Am may have slightly younger age of onset and more severe disease than Caucasians
- Age:
  - Occurs at any age, with most during childbearing years

III. Estimating SLE Prevalence in the City of Boston

Objectives for SLE Surveillance in Boston

- To develop a surveillance system using medical record abstraction from Boston & area hospitals
- Determine SLE case/patient eligibility
- To demonstrate the ability to link via a geographic information system (GIS) to existing hazardous waste site databases

SLE Partnership

- Scientific Advisory Committee
- Community Advisory Committee

Methodology

- Medical records identified for patients with SLE coded visits:

  ICD 9 code 695.4 (LE excludes S)
  ICD 9 code 710.0 (SLE)

- 11 hospital data bases (includes inpatient & outpatient visits)

Eligibility Criteria for Prevalence Study

- Resident of the city of Boston during 10/01/2003 - 9/30/2004
- One or more office visits or hospitalizations between 10/1/03 and 9/30/04

Response to HIPPA Barrier

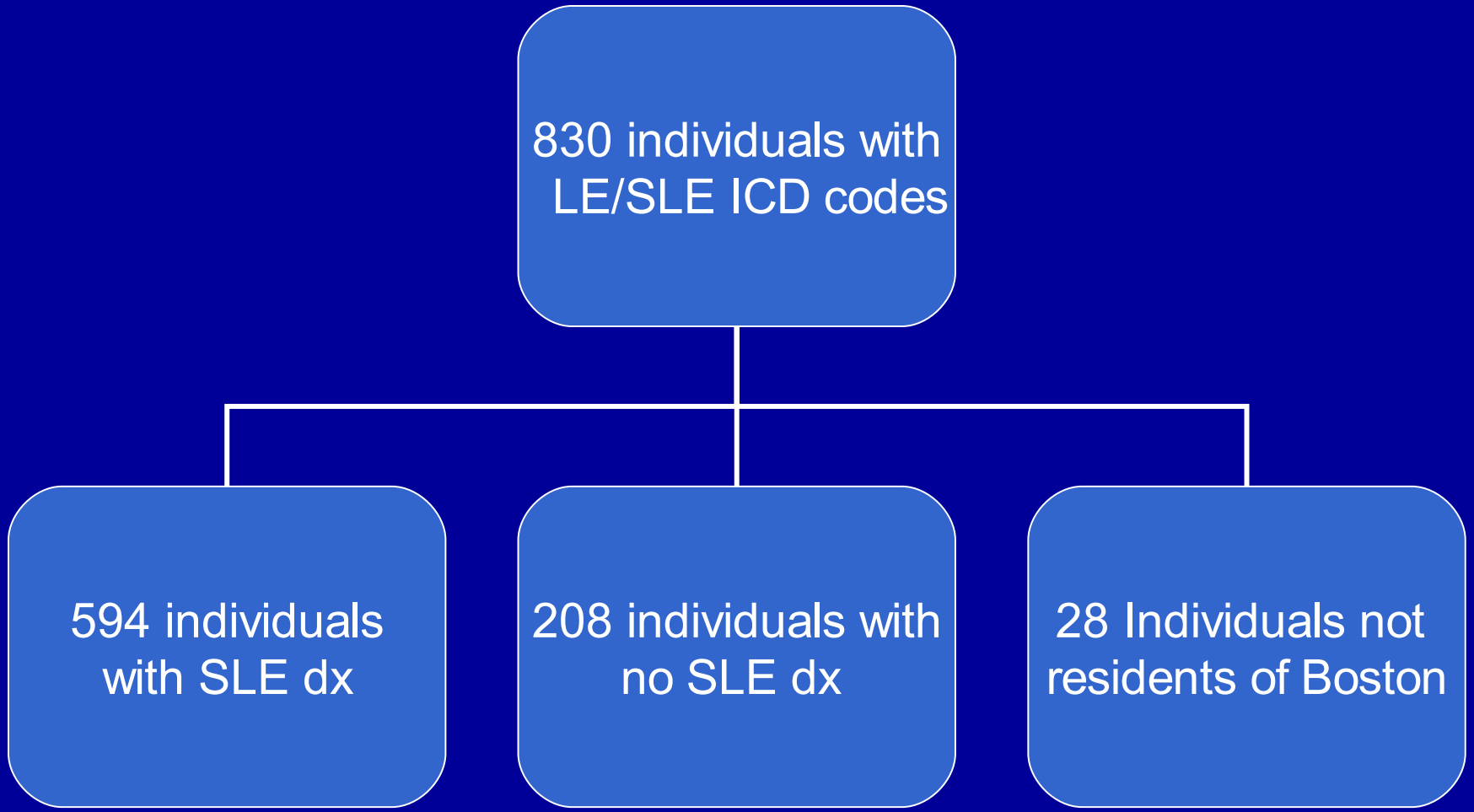
- Amended regulations that define diseases dangerous to public health with **Surveillance of Diseases Possibly linked to Environmental Exposures** (105 CMR 300.192) 2004.
- 8 diseases (ALS, aplastic anemia, asthma, autism, MS, myelodysplastic syndrome, scleroderma & SLE)

Case Ascertainment from Medical Records

927
Records Abstracted

830
Individuals with LE/SLE ICD codes

Case Ascertainment (cont.)



Prevalence of SLE

- Most literature estimates prevalence in US males at about 3/100,000 and in US females at about 49/100,000
- NHANES III self-reported physician dx: 241/100,000
- NHANES III self-reported physician dx with tx: 100/100,000
- Prevalence among black women about 400/100,000

SLE Prevalence in Boston Oct 2003 – Sept 2004

	Cases	Population	Rate per 100,000
All Races	**594**	**589,141**	**100.8**
Non-Hispanic White	183	291,561	62.8
Non-Hispanic Black	270	140,305	192.4
Hispanic	93	85,089	109.3
Other/ Unknown	48	NC	NC

Completeness: Evidence for Underestimation of SLE Prevalence

- BI Length of Surveillance Period Study/Data
- Neighborhood Health Center Survey
- Statewide Rheumatologist Survey

SLE Coded Patients Identified and Length of Surveillance*

Year(s)	months	# patients	Cum excess	increase
2003	12	76	NA	NA
2002-2003	24	98	22	29%
2001-2003	36	113	37	49%
2000-2003	48	132	56	74%
1999-2003	60	155	79	104%
1998-2003	72	185	109	143%

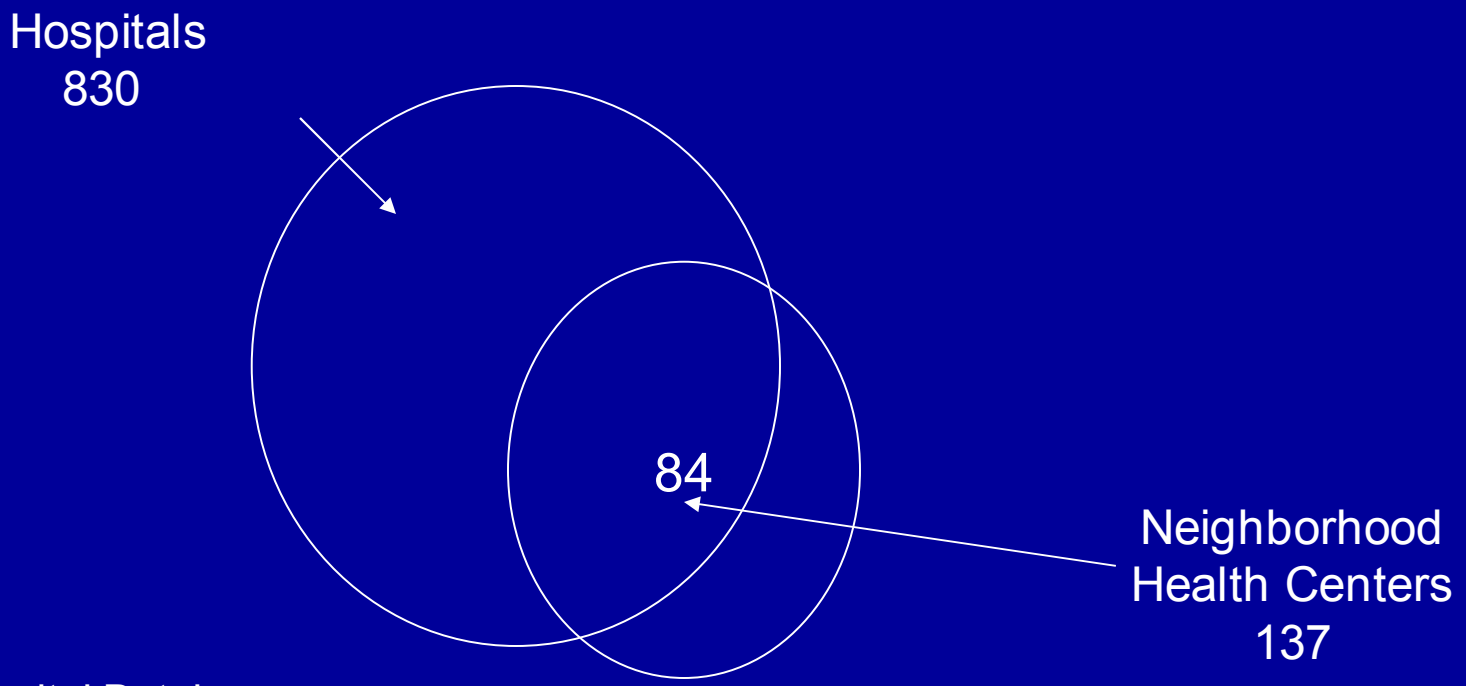
*Query of BI hospital database for unique patients with ICD-9 710.0 coded visits

Possible Explanations

- Medical follow-up obtained elsewhere (came only for consultative services)
- Inactive or mild case of SLE
- Underuse of medical care
- Relocation
- Death

Individuals Identified By Hospitals and/or Neighborhood Health Centers

(By Date of Encounter 10/1/03 – 9/30/04)

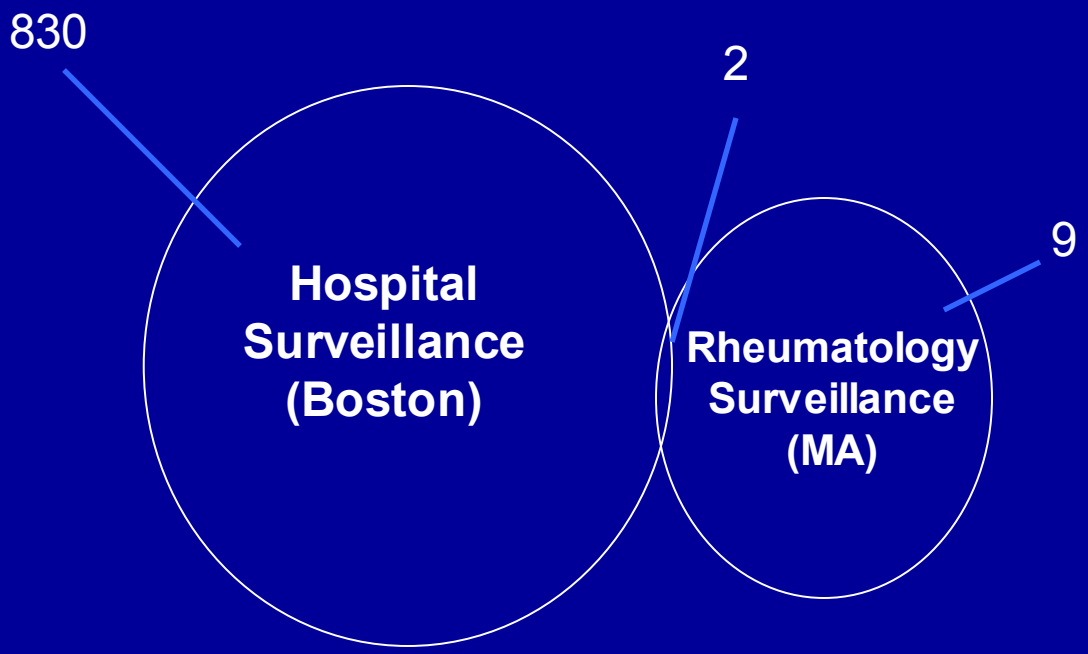


- Hospital Databases
- ICD – 9 Codes
- 710.0 (SLE) + 695.4 (LE=Non-Systematic Lupus) for 1º or 2º Diagnosis

Statewide Surveillance

- Letter to all rheumatologists (n=241) included a follow up letter 5 weeks later
- 12 letters undeliverable or not practicing in rheumatology
- 91 responded with 32 reporting 0 eligible patients

Matches for Hospital & Statewide Rheumatologist Survey



- Boston Patients Missed: 7
- Reported by Physicians in Boston: 3
- Reported by Physicians Outside Boston: 4

IV. Linkage Demonstration Project

Purpose:

- To link measures of SLE occurrence within Boston to available environmental databases
- To address feasibility and utility issues in terms of future surveillance

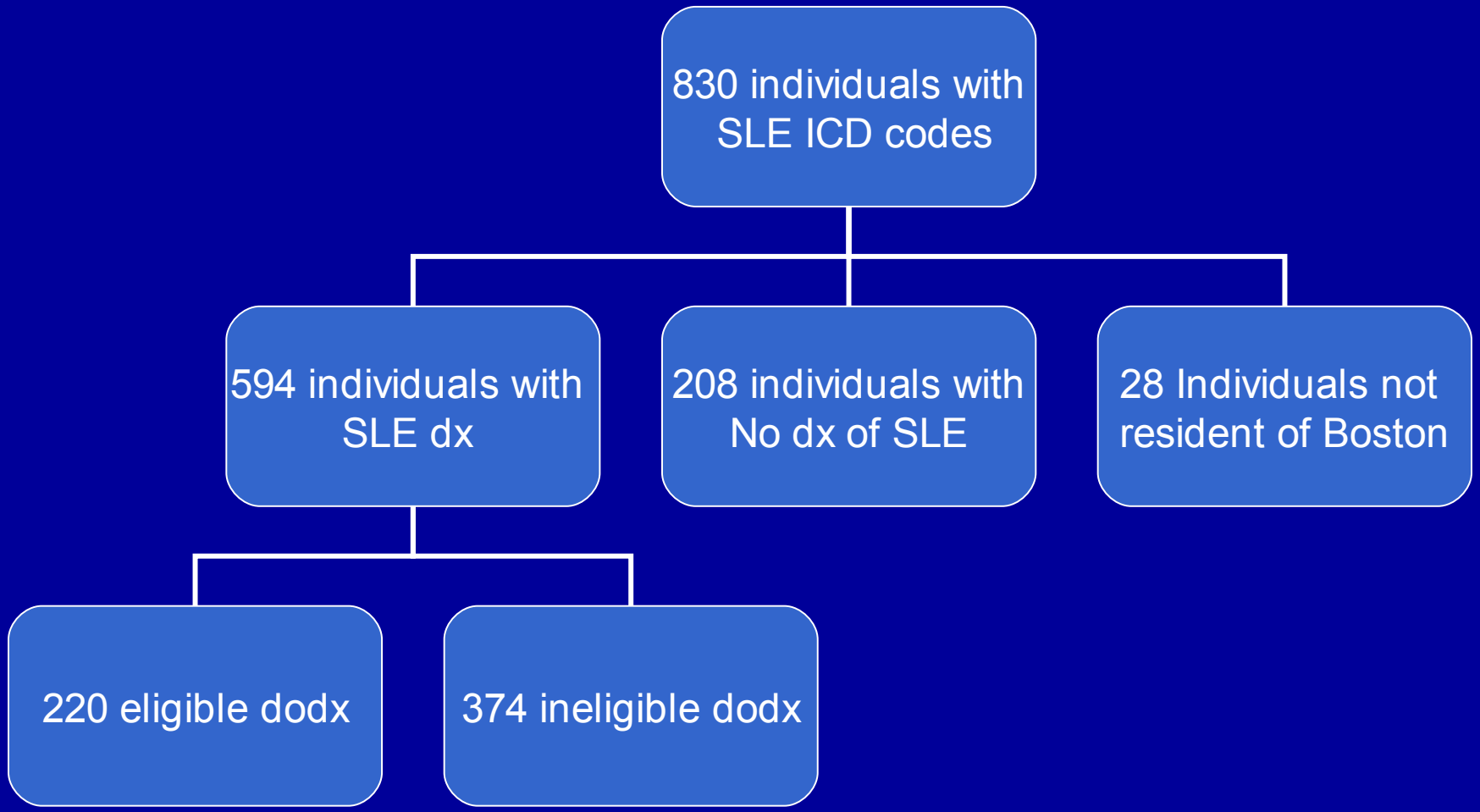
Rationale for Linkage

- Literature suggests that petroleum distillates, mercury, silica and chlorinated hydrocarbons may be associated with SLE and other undifferentiated connective tissue diseases.

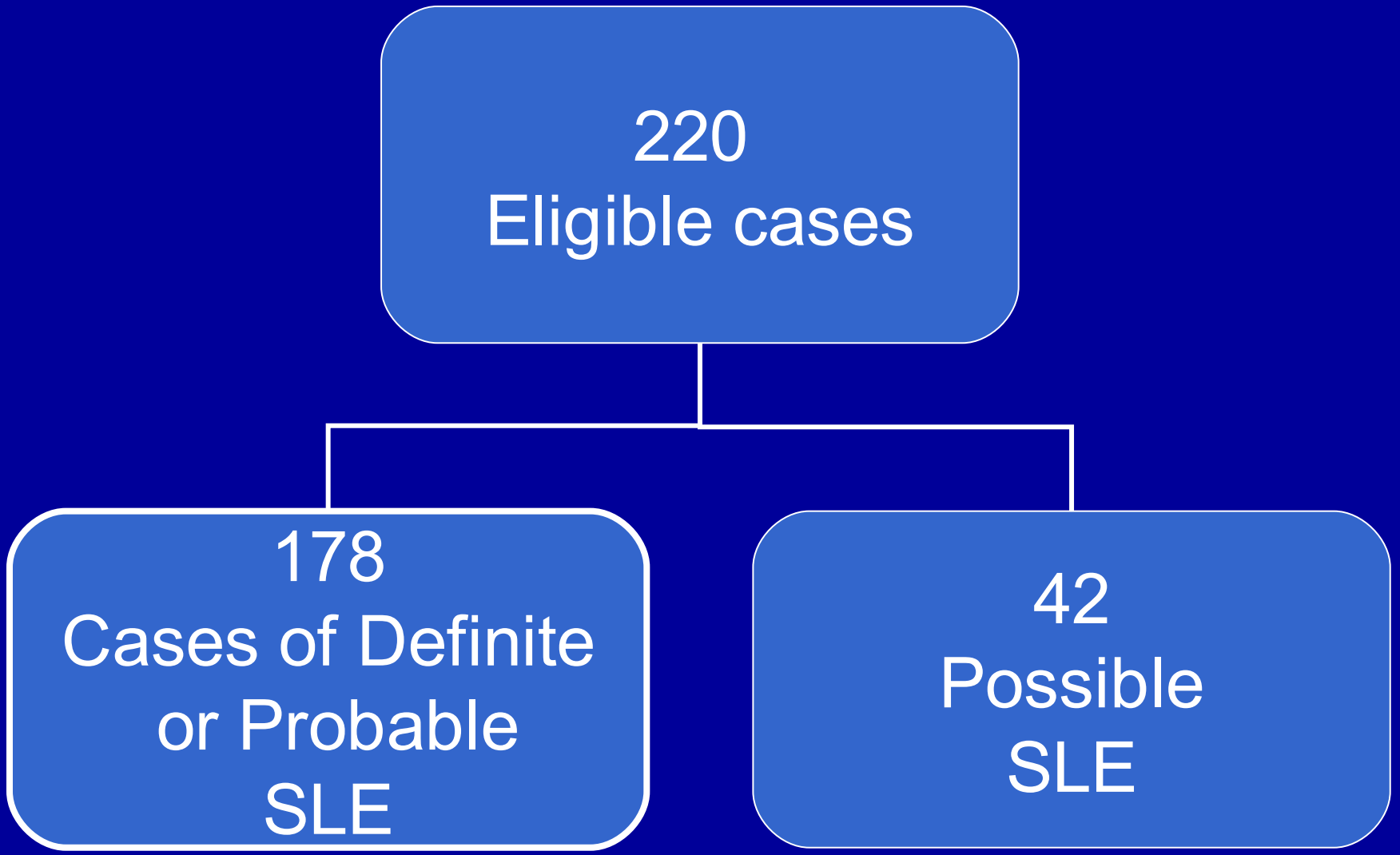
Eligibility Criteria For Linkage Study

- Resident of the city of Boston during 10/01/03 - 9/30/04
- Had at least one office visit or a hospitalization between 10/1/03 and 9/30/04
- **Diagnosed with SLE between 1/1/1999 and 9/30/2004**

Eligible Population Linkage Demonstration



Eligibility cont.



Determining SLE Diagnosis

- Language of expert's (rheumatologist, immunologist, nephrologist)?
- Date when first ACR criteria is met?
- Date when 4 out of 11 ACR criteria met?
- Date SLE first mentioned in record?
- Date of first clinical dx?
- Which date?

American College of Rheumatology Criteria

- 11 specific criteria
- Intended as a classification system for cases in clinical studies
- Individual must have 4 or more of the criteria present for the diagnosis of SLE to be made
- Documentation of 4 out of 11 criteria is considered a **definitive SLE** diagnosis

Linkage Data Available 1986-Onward

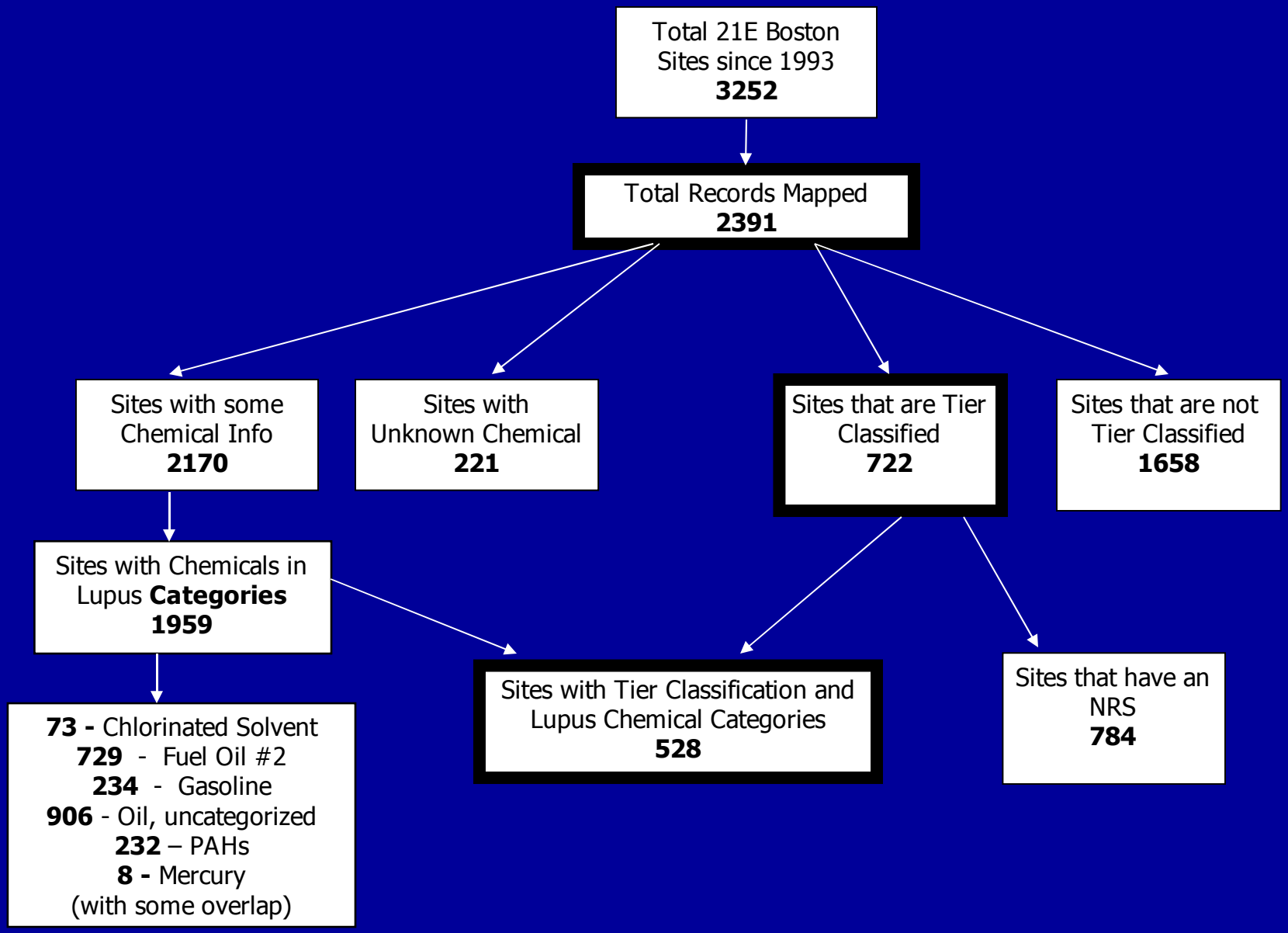
21 E sites:

- hazardous waste spills
- date
- assessment
- remedial response

Tier classified sites prioritized by:

- hazard type
- proximity to drinking water

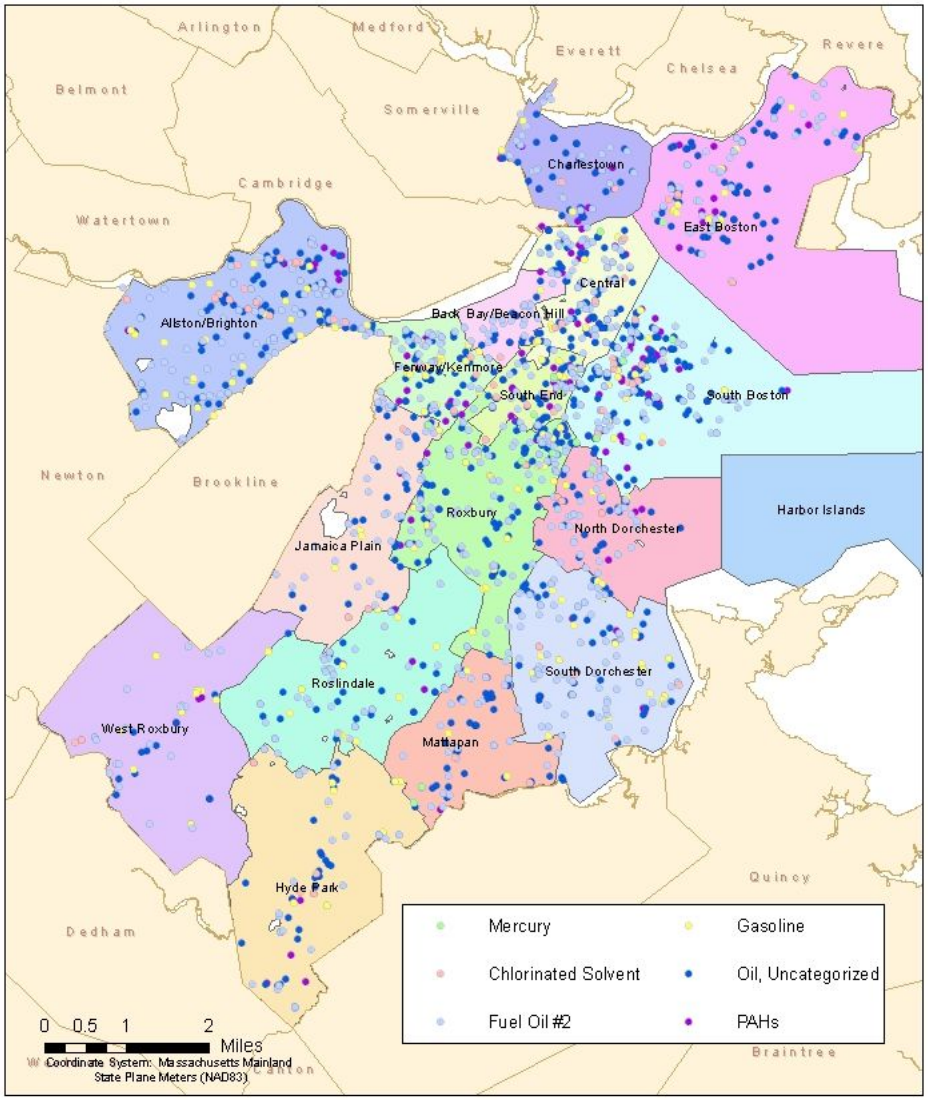
Lupus Tracking 21E Data Selected



Linkage Analysis

- Prevalence of SLE estimated by Boston Neighborhood for 1/1/99-9/30/04 diagnoses
- Density per square mile of Hazardous Waste sites by type (Tier Classified, Lupus-Suspect Contaminant sites, etc) estimated
- Statistical methods applied

Total Chapter 21e Hazardous Waste Sites by
Chemical Classification
Boston, Massachusetts



Center for CEH Environmental Health

Geographic data supplied by:
Massachusetts Executive Office of Environmental Affairs, MassGIS;
Geographic Data Technology, Inc.;

Relationship between SLE and Density of Hazardous Waste Sites (all mapped sites combined) in Boston Neighborhoods† All Races January 1, 1999 – September 30, 2004 Diagnoses

	Lupus Cases		Population	
Density of Sites†	#	%	#	%
≥ 73/sq mile	55	30.9	239,496	33.7
< 73/sq mile	123	69.1	349,645	66.3
Total	178	100.0	589,141	100.0

Chi square 7.02
P<0.01

† Based upon 2,391 sites

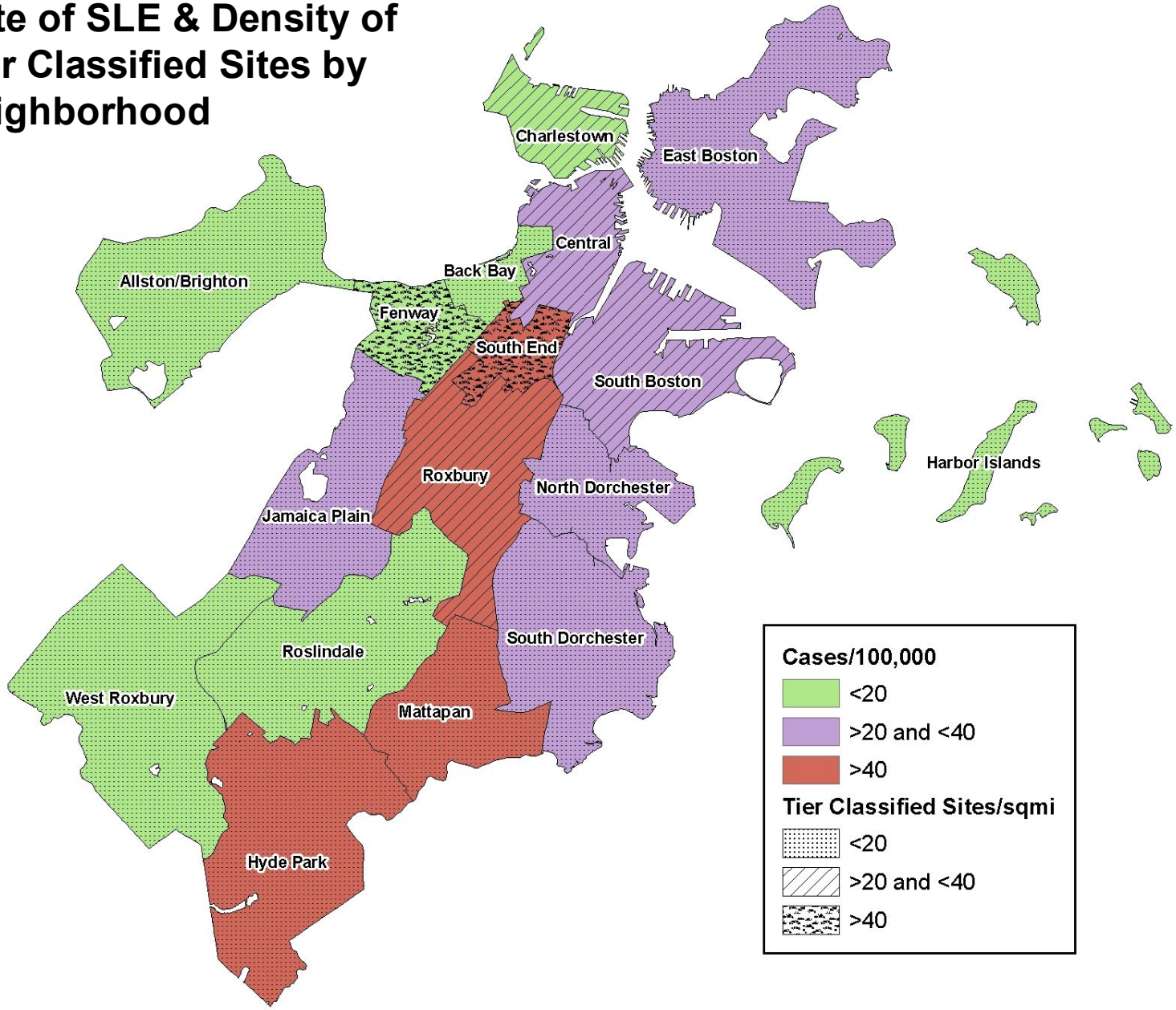
Relationship between SLE and Density of Tier Classified Hazardous Waste Sites in Boston Neighborhoods†
All Races
January 1, 1999 – September 30, 2004 Diagnoses

	Lupus Cases		Population	
Density of Sites†	#	%	#	%
≥ 29/sq mile	70	39.3	183,469	31.1
< 29/sq mile	108	60.7	405,672	68.9
Total	178	100.00	589,141	100.0

Chi square 5.56
P<0.02

† Based upon 722 sites

Rate of SLE & Density of Tier Classified Sites by Neighborhood



Relationship between SLE and Density of Hazardous Waste Sites that are Tier Classified with Lupus-Suspect Contaminants* in Boston Neighborhoods†
All Races
January 1, 1999 – September 30, 2004 Diagnoses

	Lupus Cases		Population	
Density of Sites†	#	%	#	%
≥ 15/sq mile	73	41.0	198,664	33.7
< 15/sq mile	105	59.0	390,477	66.3
Total	178	100.0	589,141	100.0

Chi square 4.23
P=0.04

* Lupus-suspect contaminants = chlorinated solvents, PAHs, mercury, oil, gasoline.

† Based upon 528 sites

Results

- Existing hazardous waste site database successfully linked by site and case geocode (to case address)
- Results are not precise because prevalence was estimated based only upon diagnoses made 1/1/99 - 9/30/04
- Summary measure for potential environmental exposure to certain types of Hazardous Waste sites were noted to be statistically significantly associated with prevalence however caution should be taken when drawing any conclusions to linkage at this point.

VI. Summary of the EPHT Experience

Lessons Learned

- Results of “most recent” DEP site analysis not readily available for surveillance.
- Environmental databases are developed primarily for regulatory purposes.
- Environmental databases contain insufficient data for meaningful linkage with health outcomes.

Lessons Learned cont'd

- Majority of physicians in private practice do not have EMRs.
- Even though physicians have electronic billing systems, these do not include variables of importance to surveillance e.g. date of diagnosis.
- Limiting analysis to one 12 month period may not be best indicator of incidence of a disease.

Lessons Learned cont'd

- Hospital medical records are not the most comprehensive place to find SLE cases.
- Retrospective record review is not the most efficient way to collect information on this disease.
- SLE diagnosis & date of diagnosis are difficult to determine in the medical record.

Recommendations

- Conduct prospective surveillance
- Survey physician/rheumatologists in all settings (private practice, clinics & health centers)
- Consider verification of each case by a rheumatologist